The Gerbode defect — a frequent echocardiographic pitfall

Ubytek typu Gerbode — pułapka dla echokadiografisty

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Abstract

We present a case of Gerbode type defect (left ventricular to right atrial communication) discovered in a 52 year-old man with atrial and ventricular septal defects. The patient was diagnosed using two-dimensional colour Doppler and transoeso-phageal echocardiography. We describe the echocardiographic features and review the anatomical consequences of such defects. Our aim was to remind readers about this rare organic heart disease that an inexperienced echocardiography specialist might easily mistake for a recoil wave of tricuspid valve incompetence and thus diagnose pulmonary hypertension.

Key words: left ventricular to right atrial shunt, congenital heart defects, transoesophageal echocardiography

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Left ventricular-to-right atrial communications are rare types of ventricular septal defect (VSD) known as the Gerbode defect.

A 52 year-old man was referred to cardiologic admissions by his general practitioner because of arterial hypertension and holosystolic heart murmur. Medical history taken from the patient revealed that he had had a headache and buzzing noise in his head for five days. His blood pressure taken on an outpatient basis reached up to 180/100 mm Hg, and his effort tolerance was good. The patient had not taken any medications chronically and had not previously been monitored systematically.

Major deviations were found — a loud holosystolic murmur (5/6 on the Levin scale) and a murmur in the region of the apex of the heart and left margin of the sternum attracted special attention. Blood tests revealed high values of transaminases. Morphology, ionogram, creatinine, glucose and clotting times were correct.

Chest X-ray revealed interstitial and central haemostasis in the lungs, considerable enlargement of the left heart, and aorta with atheromatous changes.

Electrocardiogram revealed steady sinus rhythm with a 75/min rate, nomogram, incomplete right bundle branch block, and left-ventricular hypertrophy.

Echocardiography enabled a diagnosis of enlargement of the left atrium (46 mm), and of a not enlarged left ventricle (52/34) with thickened muscle (15 mm). Left ventricle contracted correctly, ejection fraction amounted to 60%. Mitral valve and aortic valve did not display any organic changes, although a minor/moderate aortic incompetence was observed. Right cardiac chambers were enlarged — right atrium 50 mm, right ventricle 46 mm. In the right atrium, a systolic flow from the attachment of the tricuspid valve's septal leaflet with gradient up to 140 mm Hg was observed, which was initially misinterpreted as tricuspid incompetence (Figs. 1, 2) (and indeed turned out to be a left ventricle-to-right atrial shunt). The acceleration of flow in the pulmonary artery amounted to 120 ms. Pulmonary artery was enlarged. In addition, echocardiography revealed an aneurysm of the membranous part of ventricular septum and a defect 5 × 6 mm in diameter

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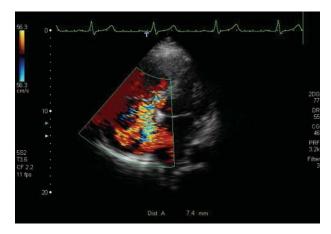


Figure 1. Transthoracic four-chamber view with systolic flow initially misinterpreted as tricuspid incompetence



Figure 3. Midoesophageal four-chamber view showing a jet from the left ventricle into right ventricle (ventricular septal defect) and from the left atrium into right atrium (atrial septal defect)

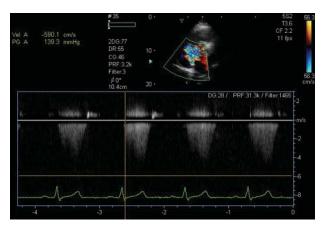


Figure 2. Transthoracic four-chamber view with systolic gradient estimated in the right atrium originating from the membranous septum

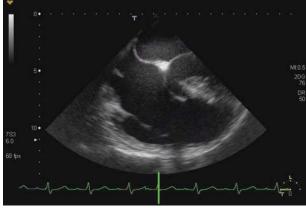


Figure 4. Midoesophageal four-chamber view with ventricular septum aneurysm

with left-to-right shunt and gradient up to 130 mm Hg. Qp/Qs amounted to about 3.0.

Transoesophageal echocardiography was conducted. This examination disclosed enlarged right cardiac chambers and a defect in atrial septum (8 mm) in a typical place with left-to-right shunt (2–3 mm margin from aorta, 7–11 mm from posterior wall). Furthermore, the examination also revealed an aneurysm in the membranous part of ventricular septum with left ventricle-to-right ventricle shunt 3.5 mm wide (Doppler marked with colour) and gradient up to 120 mm Hg (Fig. 3). Having made a thorough assessment of the atrioventricular septum, it was also possible to display an aneurysm (Fig. 4) with left ventricle-to-right atrial shunt (Fig. 5) 8.6 mm wide and gradient up to 140 mm Hg. On the basis of both transthoracic and transoesophageal echo-



Figure 5. Midoesophageal four-chamber view with colour-flow Doppler

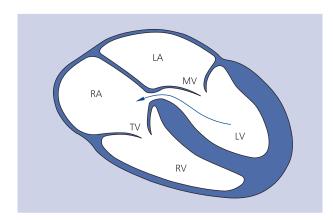


Figure 6. Gerbode defect scheme; RA — right atrium; LA — left atrium; LV — left ventricle; RV — right ventricle; MV — mitral valve; TV — tricuspid valve

cardiography, atrial septal defect of ASD II-type, defect in the membranous part of ventricular septum, and Gerbode--type defect were diagnosed.

Gerbode-type defect is a left ventricle-to-right atrial shunt (Fig. 6). In 1958, Gerbode was the first to describe several cases of surgical closing of this defect. It is considered a very rare type of VSD. Its occurrence is expressed by the following ratio: 8 cases to 10,000 of congenital diseases. There are certain secondary causes of the defect under discussion, including a history of endocarditis, chest injuries, cardiac infarction, and condition after the replacement of a mitral or aortic valve. It is also believed that a number of Gerbode-type defects are secondary to a congenital VSD-type defect — there

are several natural ways of closing the VSD, one of which is the formation of an aneurysm via hyperplasia of connective tissue covering the leaflets of tricuspid valve that touch the margin of the defect. There are two kinds of Gerbode-type defect, namely supravalvular and subvalvular. Supravalvular defect covers the atrial septum and is found just over the attachment of the septal leaflet of the tricuspid valve (artioventricular part of membranous septum). Subvalvular defect — which is more common — covers the membranous part of ventricular septum and is always connected with the defect of septal leaflet of tricuspid valve.

Our aim was to remind readers about this rare organic heart disease that an inexperienced echocardiography specialist might easily mistake for a recoil wave of tricuspid valve incompetence and thus diagnose pulmonary hypertension.

Conflict of interest: none declared

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